EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 5
Issue: 11
24-26
Year: 2018

DROP METASTASIS TO THE THORACIC SPINAL CORD FROM A JUGULO-TYMPANIC PARAGANGLIOMA WITH INTRA-CRANIAL EXTENSION. CASE REPORT AND LITERATURE REVIEW

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Article Received on 24/09/2018 **Article Revised on 13/09/2018** **Article Accepted on 04/10/2018**

ABSTRACT
Cerebrospinal fluid dissemination (CSF) of paragangliomas has been rarely reported and only with spinal primaries. Time to cerebrospinal fluid dissemination is highly variable thus optimal management remains to be determined. A literature review identified four males and two females who presented with CSF dissemination following management of a spinal paraganglioma. One female patient who presented with drop metastasis from a jugulotympanic paraganglioma was also included. Mean age was 41 years (ranged from 15 to 74). Time to CSF dissemination ranged from four months to 22 years and multiple metastases were present in three patients. Metastasis management included surgery in two patients, surgery and adjuvant radiotherapy in two patients, radiotherapy alone in two patients and in one patient with multiple metastases peptide receptor radionucleid therapy with radiolabeled somatostatin analog was employed. Following treatment three patients were reported to improve, two worsened, and two remained stable. Long-term follow-up of patients diagnosed with intradural paragangliomas is necessary. Prospective studies are required to define optimal management of central nervous system paragangliomas presenting with CSF seeding.

KEYWORDS: Paraganglioma; Jugulo-tympanic; Metastasis; Spinal cord; Intradural; Cerebrospinal fluid dissemination.

INTRODUCTION
Paraganglioma is defined by the World Health Organization as "a unique neuroendocrine neoplasm, usually encapsulated and benign, arising in specialized neural crest cells associated with segmental or collateral autonomic ganglia (paraganglia); consisting of uniform chief cells exhibiting neuronal differentiation forming compact nests (Zellballen) surrounded by sustentacular cells and a delicate capillary network"[1] Head and neck paragangliomas constitute 0, 6% of head and neck tumors and 0, 03% of all tumors.[1] Jugulo-tympanic paragangliomas most commonly occur in the fifth and sixth decade of life and demonstrate female predilection. Extension into the intracranial cavity occurs in 36% of the cases and manifests with pulsatile tinnitus and lower cranial nerve palsies.[1] Malignancy is defined by the presence of distant metastasis[1,2] and has been reported to occur in 2-4% of jugulo-tympanic paragangliomas.[2]

Cerebrospinal fluid (CSF) dissemination of paragangliomas has been rarely described[3-8] thus, optimal patient management remains unclear.

We report on a 74-year-old female who presented with a late, thoracic spinal cord, drop metastasis from a previously treated right jugulotympanic paraganglioma and review the relevant literature for this unusual condition.

CASE REPORT
A 74-year-old female presented to the neurosurgery clinic with a two-week history of progressive bilateral lower limb weakness. On neurological examination she had spastic paraparesis and hypoesthesia localized approximately at the T4 level. Babinski sign was present bilaterally. According to the patient, 10 years prior to presentation she underwent resection of a right jugulo-tympanic paraganglioma. Six years following operation recurrence was noted on follow-up Magnetic Resonance Image (MRI) (fig 1A) which was managed with radiosurgery (Cyberknife 2700 cGy). Following radiosurgery she suffered from dysphonia and severe dysphagia necessitating percutaneous gastrostomy placement.

During current admission, thoracic spine MRI was obtained which demonstrated a posterior, intradural, extra-medullary, contrast enhancing, space occupying lesion compressing the spinal cord at the T1-T2 level (fig 1B). The patient underwent T1 and T2 laminectomy aiming for total tumor resection. However, due to severe intra-operative bleeding and hemodynamic instability partial resection of the tumor was done (fig 1C). Histopathology revealed paraganglioma (fig2a-f). Post-operatively, improvement of her symptoms was noted. On postoperative day seven she was referred for radiotherapy. On the one-month follow-up she was able to walk with a cane.

Figure 1:

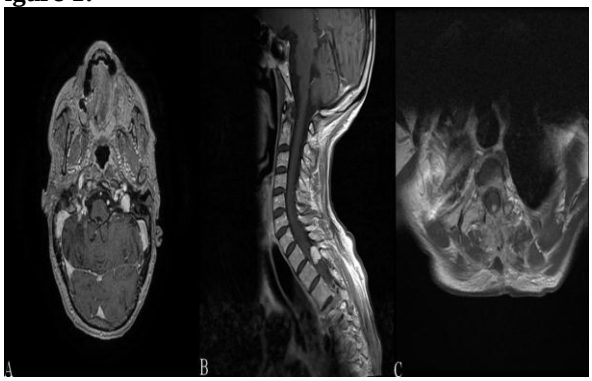

Figure 1: Figure 1: (A) Post-radiosurgery, axial, T1 Weighted, Magnetic Resonance Image (MRI) demonstrating intracranial extension of the paraganglioma (B) Pre-operative, T2 Weighted MRI, saggital demonstrating an intradural, extramedullary, uniformly contrast enhancing lesion. (B) Post-operative, axial, T2 Weighted contrast enhancing spinal MRI significant for tumor remnant.

Figure 2:

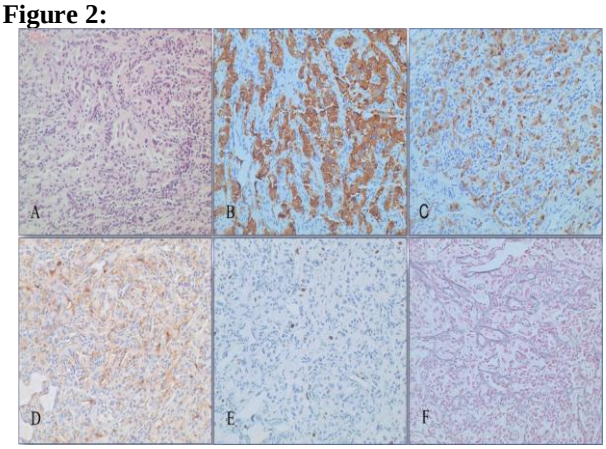

Figure 2: (A) The tumour is composed of round-to-oval large cells with large nuclei and abundant granular eosinophilic cytoplasm arranged in nests (zellballen) within a prominent vascular network (Hematoxylin–Eosin x200). (B and C) On immunohistochemical staining the tumour cells demonstrate expression of neuro-endocrine markers (Chromogranin x200 and Synaptophysin x200), whereas S100p (D) is expressed by sustentacular cells (S100p x200). (E) The mitotic index was approximately 5% (Ki-67 x200). (F) Reticulin histochemical stain highlights the Zellballen pattern (Reticulin x200).

Literature Review

Six single-patient case reports describing CSF dissemination of spinal paragangliomas were identified.

Including the current case there were four males and three females. Patient age ranged from 15 to 74 years old (mean age 41 years). The primary tumor was located in the cervical spine (n=1)[3], the thoracic spine (n=1)[4], the lumbar spine (n=4)[5-8] and the right Jugulotympanic region extending intracranially (n=1). Management of the primary tumor included total resection and adjuvant radiotherapy (n=2)[5,6], total resection alone (n=1)[7], partial resection and adjuvant radiotherapy (n=2)[3,8] and partial resection without radiotherapy (n=2).[4] Time from diagnosis of paraganglioma to CSF dissemination ranged from four months[4,6] to 22 years.[7]

Multiple metastases were present in three patients[3,6,8] and single metastasis in four patients.[4,5,7] Metastasis management included surgery in two patients[5,7], surgery and adjuvant radiotherapy in two patients[8], radiotherapy alone in two patients[3,4] and in one patient with multiple metastases peptide receptor radionucleid therapy with radiolabeled somatostatin analog was employed.[6] Following treatment of the metastases three patients were reported to improve[5,6], two worsened[7,8] and two remained stable.[3,4]

DISCUSSION

Paraganglioma CSF dissemination has been rarely reported. Including the case presented here a total of seven patients have been reported. The exact mechanism responsible for CSF dissemination remains unknown. Factors proposed to be associated with paraganglioma spread via the CSF include: young age at diagnosis[7], partial versus total resection of the primary tumor[5] and failure to perform extracapsular resection of the primary tumor.[5,7]

Time from diagnosis to CSF dissemination is highly variable. In our review it ranged from four months[4,6] to 22 years[7], indicating the necessity for very long-term follow-up of patients diagnosed with intradural paraganglioma.

Optimal management of paraganglioma CSF metastasis remains to be determined. Surgical resection alone may be offered to all cases of accessible lesions for which gross total resection may be possible, and in cases of symptomatic or life-threatening lesions. Radiotherapy should be recommended as an adjuvant treatment following partial resection of the metastasis. In cases of multiple intradural metastases not amenable to surgical management, radiotherapy or peptide receptor radionucleid therapy with rediolabeled somatostatin analog may be considered.

In conclusion, we report on a rare case of a late, thoracic spinal cord drop metastasis from a jugulotympanic paraganglioma with intracranial extension. Our case indicates the importance of long-term clinical and imaging follow-up of patients diagnosed with central nervous system paragangliomas.

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